Radiology Corner

Cystic Fibrosis Chest X-Ray Findings: A Teaching Analog

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Note: This is the full text version of the radiology corner question published in the June 2008 issue, with the abbreviated answer in the July 2008 issue.

The abnormal chest radiograph is often perplexing, but abnormal findings can be interpreted easily with a systematic and methodical approach. The companion cases presented this month highlights two basic, but often forgotten or confused findings that have very different meanings: tram-tracks and air-bronchograms. The authors present these two cases along with the assistance of analog imaging phantoms constructed out of household materials (straw, sponge, tape and apple), and application of some contrast media. The authors believe the presented representative cases of bronchial wall thickening (in the CF case highlighted here) and the finding of tram tracks, compared to consolidation with typical air-bronchograms, should make these findings conceptually clear. An organized differential accompanies each radiographic finding to complement these two specific known diagnoses.

Introduction

There are countless radiographic "signs" and eponyms involving the common chest x-ray. While many are very specific for a certain disease and others are quite rare, there are a few findings that could be seen on any given day in a busy hospital. The purpose of this article is to provide a clear model for distinguishing two common but key findings, and providing a proper differential diagnosis based on that distinction. Both "tram-tracks" and "air bronchograms" indicate that the airways, normally not visible amongst the lung tissue, can often be traced. The key to this phenomenon is that the pathology for each process is unique, and leads to a separate differential.

Historically, the "tram-track sign" has been used as a descriptive radiographic term. For example, there is "tram-tracking" of the optic nerve meninges that can indicate a meningioma [1]. Neurology employs a "tram-track" sign that indicates cortical calcifications as seen in patients with Sturge-Weber Syndrome [2]. Regardless of the source, "Tram Tracks" are parallel linear densities that stand out from their surrounding tissues. The version discussed below is that of pulmonary tram-tracking, which indicates the visibility of thickened bronchial walls that do not show normal tapering toward the lung periphery.

Air-bronchograms, by comparison, do not involve any thickening or inflammation of the bronchioles. Historically, the term "bronchogram" was used to describe a radiograph taken after the inhalation of a radio-opaque substance such as barium sulfate [3]. This would provide an impressive picture of the bronchiole architecture and was used for exploring pulmonary processes prior to the advent of CT imaging. Currently, the commonly used term "air bronchogram" has implication that the "contrast material" is the normal air within the bronchioles. In contrast to the original bronchogram, the bronchioles are only visible when a pathologic process involving the *surrounding tissue* creates a significant change from the normal lung density and highlights the lucency of the air-filled bronchioles.

To illustrate the differences between tram-tracks and air-bronchograms, an experimental model was created. Household items including kitchen sponges soaked in both tap water and IV contrast material, an apple, and straws of various diameter were imaged using both plain film x-rays as well as a CT scanner. The water-soaked sponges provide the baseline "lung tissue," allowing visualizing a portion of the architecture, but with most of the image being radiolucent. Straws placed into the sponges had varying appearances depending on the diameter of the straw and the thickness of the straw wall. To create "bronchiole thickening" it was necessary to wrap the straws with several layers of medical tape soaked with contrast. An apple was used in place of the sponge to simulate lung tissue that was consolidated. Straws pushed through the apple appeared as tubular lucencies when contrasted to the dense surrounding tissue, and there was no appreciable visualization of the straw itself. These models will provide the analog images used in this article.

Summary of Imaging Findings

The images shown in Fig 1(A) and 1(B) are the cases that exemplify the tram-track sign (A) and the Air-Bronchogram (B). The case images and descriptions are followed by teaching analogs with our experimental models.

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Report Documentation Page

Form Approved
OMB No. 0704-0188

1. REPORT DATE	2. REPORT TYPE	3. DATES COVERED
JUL 2008		**00-00-2008 to 00-00-2008**

4. TITLE AND SUBTITLE **Cystic Fibrosis Chest X-Ray Findings: A Teaching Analog**	5a. CONTRACT NUMBER 5b. GRANT NUMBER 5c. PROGRAM ELEMENT NUMBER
6. AUTHOR(S)	5d. PROJECT NUMBER 5e. TASK NUMBER 5f. WORK UNIT NUMBER
7. PERFORMING ORGANIZATION NAME(S) AND ADDRESS(ES) **Uniformed Services University,Bethesda,MD,20814-4799**	8. PERFORMING ORGANIZATION REPORT NUMBER
9. SPONSORING/MONITORING AGENCY NAME(S) AND ADDRESS(ES)	10. SPONSOR/MONITOR'S ACRONYM(S) 11. SPONSOR/MONITOR'S REPORT NUMBER(S)

12. DISTRIBUTION/AVAILABILITY STATEMENT
Approved for public release; distribution unlimited

13. SUPPLEMENTARY NOTES

14. ABSTRACT

15. SUBJECT TERMS

16. SECURITY CLASSIFICATION OF:			17. LIMITATION OF ABSTRACT	18. NUMBER OF PAGES	19a. NAME OF RESPONSIBLE PERSON
a. REPORT	b. ABSTRACT	c. THIS PAGE			
unclassified	**unclassified**	**unclassified**	**Same as Report (SAR)**	**6**	

Standard Form 298 (Rev. 8-98)
Prescribed by ANSI Std Z39-18

Fig. 1 (A) PA Chest X-Ray of the cystic fibrosis patient showing multiple tram-tracks diffusely and bilaterally. Most notably: one extending laterally from the right hilum; one each extending superiorly from both hila; one extending inferior and laterally from the left hilum. Figure 4A better demonstrates this with close up of RUL with arrows pointing out the tram tracks.

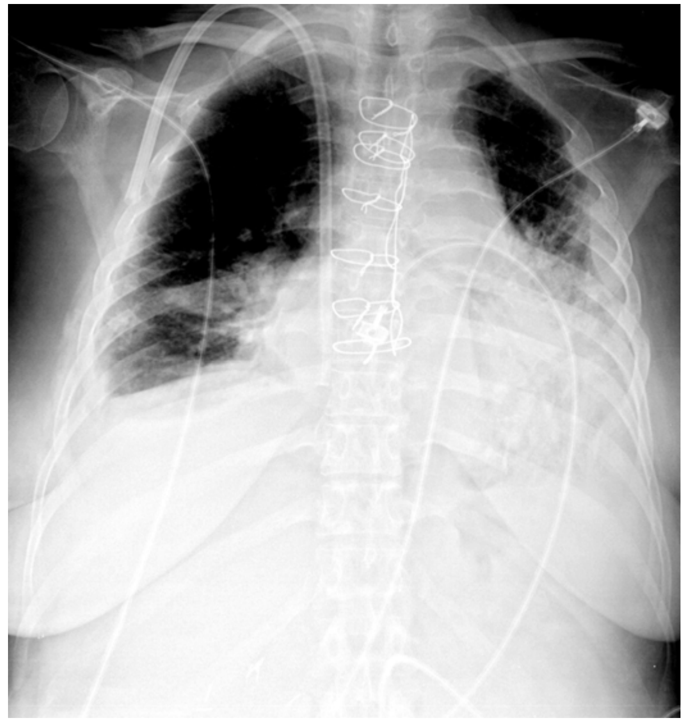

Fig. 1 (B) PA Chest X-Ray of a 53 year old post-operative woman who displays Air Bronchograms bilaterally. The left lower lung field contains a "complete air bronchogram," meaning the tubular lucency extends from hilum to periphery. The right lower lung shows a less obvious air bronchogram exiting the hilum inferiorly. Figure 4B better demonstrates this with close up of LLL with arrows pointing to the air bronchograms.

Case 1: Cystic Fibrosis: Tram Tracks

The first case highlighted as the unknown image involves a patient with cystic fibrosis, a well-known entity that involves mucus plugging of the airways, chronic bronchiole inflammation and infection, and ultimately diffuse bronchiectasis. As will be shown below, bronchiectasis is one of the more common entities presenting with pulmonary tram-tracking.

Figure 1(A) shows multiple radiographic findings consistent with the pathophysiology of cystic fibrosis. Mucus plugging of the small bronchioles results in hyperinflation of the lungs. There are multiple nodular opacities which represent the mucus plugs. The aforementioned tram-tracking is visible in all lung fields, most conspicuous in the bilateral upper lung fields and extending laterally from the right hilum. In addition, multiple ring-shaped shadows can be seen scattered throughout. This patient is a long-time sufferer of cystic fibrosis, and represents the example of a radiograph with tram-tracking secondary to bronchiectasis. As will be addressed later, this is only one possible cause of tram-track sign.

Case 2: Post-Operative Atelectasis: Air Bronchogram

A second case for comparison purposes is presented and is a 53 year old woman 2 days status-post thoracotomy who developed shortness of breath. This will be used as the air-bronchogram example. As the differential diagnosis for air-bronchograms is extensive, only superficial coverage of the disease entities mentioned will be provided.

Figure 1(B) of our companion case shows diffuse, bilateral lower lung field densities. The left heart border is obscured, as are both costophrenic angles and hemi-diaphragms. The sternal wires, ECG leads, and surgical drain are visible, and are consistent with the patient's history as a post-operative patient. Of the greatest importance for this discussion is the presence of the complete air-bronchogram in the left lower lung field and the partial air-bronchogram in the right lower lung field. The complete air-bronchogram can be seen to extend from the hilum out into the peripheral lung field, and outlines several branches of the bronchial tree. This case most likely represents atelectasis due to prolonged immobilization. Again, the possible differential is much more extensive and will be discussed in detail later.

Figure 2A and B are two photographs of the experimental setup used for the analog radiographic images. Two sponges, one half soaked with contrast, the other soaked in plain water are used for surrounding lung. Since the sponge did not soak up enough to represent airspace disease, the authors used an apple as a "hyperdense" lung, i.e. one that has undergone consolidation or atelectasis. The straws are either clear or red, red being slightly smaller diameter (mentioned for reference). All straws used in this project are hollow unless otherwise noted. The upper image (Figure 2A) shows the plain film X-ray cartridge used, and the lower image (Figure 2B) is taken on the CT scanner bed, resting on a towel.

Plain film and CT of the models in Fig 2 (A,B) are presented to illustrate a clear conceptualization of the pertinent signs. Figure 3(A) and 3(B) are the plain film and CT images that illustrate tram-tracks and air bronchograms, respectively.

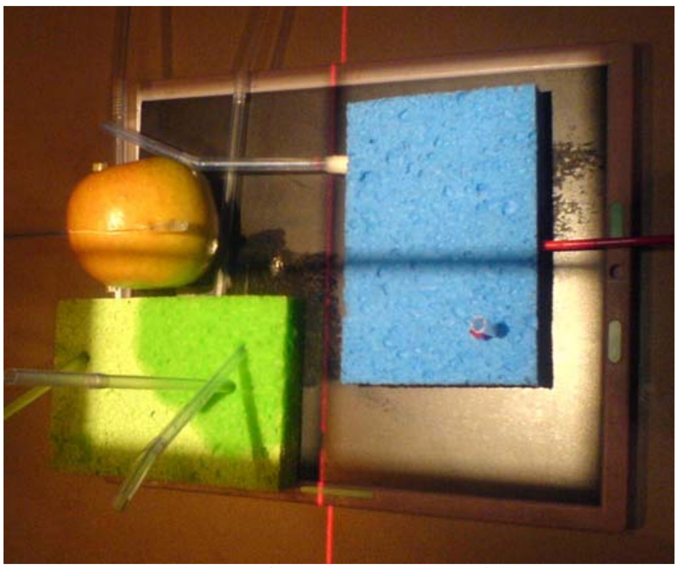

Fig. 2A. Gross photo of experimental setup for Plain Computed Radiography. The green sponge has contrast medium on half, and the blue has been soaked with water.

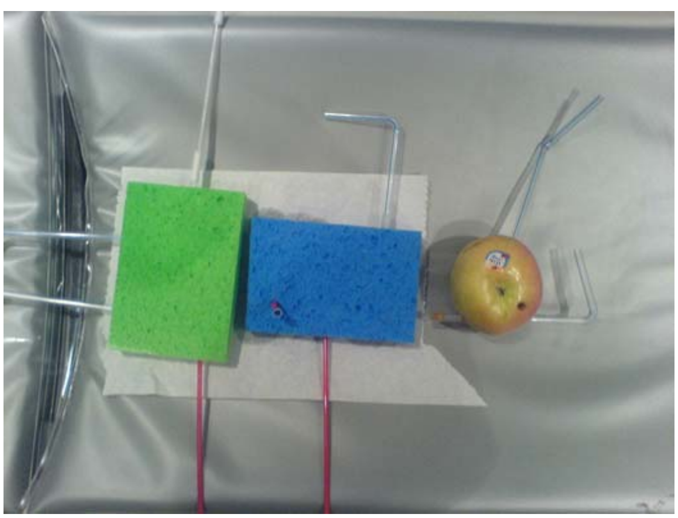

Fig. 2 B: Gross photo of experimental setup for CT. Note the red and clear straws, whole apple with straws.

Figure 3(A), the tram-track model, illustrates several features of this phenomenon. The plain film on the left shows a small diameter straw in profile as well as a larger straw in the oblique. The linear edges at the lateral margins of the straw can be seen on both. The oblique straw demonstrates the importance of thinking 3-dimensionally. The ring shadow at the upper end of the tram track is simply the point where the straw was cut, showing that ring shadows and tram-tracks are just different 2-dimensional perspectives on the same structural feature.

Figure 3(B) demonstrates the important features of an air-bronchogram. The "airway" (or straw in our model) is only visible as the product of the two edges created by the adjacent higher density apple material. The airway, in both this model and a real patient, is completely patent. This can be seen in both the end-on views and the CT image showing air throughout the lumen of the straw.

Figure 4 (A) shows the side-by-side comparison of a tram-track in the patient with CF along side the CT image of the model of the same. Likewise, figure 4 (B) compares the cropped and enlarged image of an air bronchogram in our ICU patient along side the plain film of the apple model.

Figure 5 demonstrates the final piece of the experimental model. The upper image is an axial CT of the chest of a patient with bronchiectasis. Of note in the right lung are two examples of signet rings, shown up close in the lower left image. The enlarged bronchiole is a circular ring of high attenuation and central clearing, and it is abutted by the normal sized pulmonary arteriole that tracks with it. This is abnormal, as usually the bronchiole and arteriole are approximately the same diameter. The right lower picture shows the CT model version of this phenomenon, with a ring-shaped attenuation that is a large straw on end, abutting a smaller straw which was filled with radio-opaque material to represent the arteriole.

Discussion

A thorough understanding of the mechanisms involved in creating either the tram-track sign or air-bronchogram leads naturally to creating a reasonable general differential diagnosis. Often times, the clinical features combined with the radiographic findings can lead to a fairly specific diagnosis.

Tram-tracking was presented in the experimental model as a thick-walled tube in a normal surrounding. How does this situation evolve in a patient? To create a biologic tram-track, two processes need to occur. The first is bronchiole dilation, and the second is bronchiole inflammation. Bronchiole dilation occurs readily when obstruction of the airways leads to air-trapping and mechanical stretching of the airway. Likewise, weakening of the muscular layers of the bronchioles leads to an unbalancing of the forces that maintain the airway. The outward pull of the elastic lung parenchyma causes unopposed bronchiole dilation. [5] The inflammatory component can be either acute or chronic. Acute inflammation, as in any other area of the body, leads to edema of the bronchiole. Chronic or recurrent inflammation can have a more lasting effect, causing permanent damage to the muscular and elastic layers of the airway. Additionally, damage to the ciliary layer predisposes to further obstruction and infection, setting the stage for a worsening cycle of lung injury.

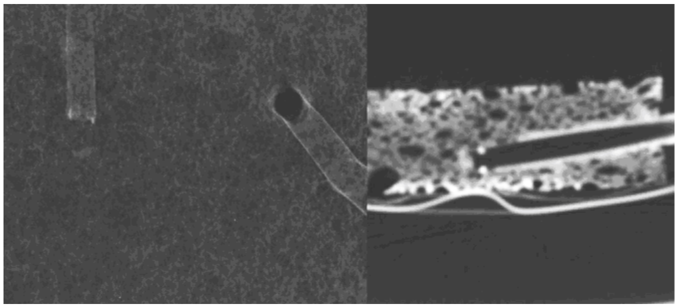

Fig. 3 (A) Tram-tracking in the experimental model, with plain film to the left, and axial CT on the right. The plain film shows one straw in profile (left) and one straw in oblique (right). The CT image shows the increased attenuation of the tape-wrapped straw mimicking bronchial wall thickening.

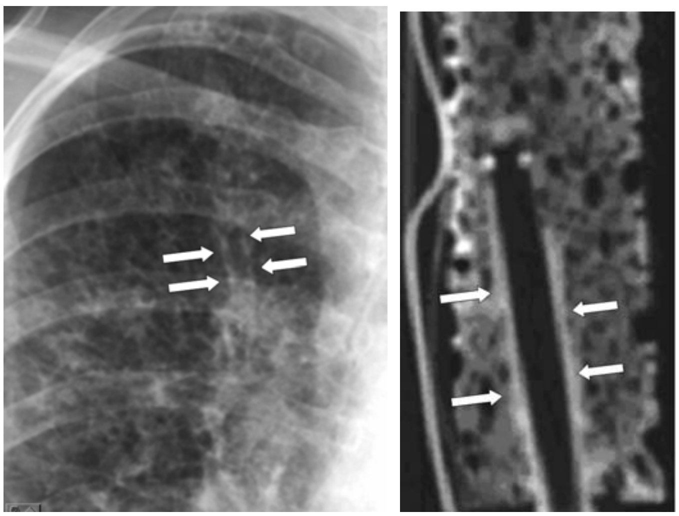

Fig. 4 (A) Side-by-side comparison of the *in*-vivo Tram-Track sign on the left and the experimental analog on the right. The arrows point to the thickened bronchiole walls seen in the CF patient, created in the same fashion by a straw wrapped in tape producing a thick wall around a hollow tube inside the sponge.

Table 1 shows the general differential for the causes of tram-tracking. This list can be broken down in several ways: Acute or Chronic, Acquired or Congenital, or by initial event (obstruction or dilation).

Acute bronchitis and bronchiolitis are both infectious processes that cause airway inflammation leading to obstruction and dilation. While this can produce the tram-tracking sign on x-ray, the pathology is generally reversible and will resolve with treatment of the underlying cause [4].

Table 1: Causes of Tram-Track Sign

Entity		Etiology
Acute Bronchitis		Generally Viral or Bacterial
COPD	Chronic Bronchitis	Secondary to prolonged tobacco use, Related to airway dilation and increased mucus production.
	Emphysema	
Bronchiolitis		Most common in infants due to RSV
Bronchiectasis		Multiple congenital and acquired causes.

Table. 1 The most common disease processes which can present radiographically with "tram-tracks" on x-ray or CT are presented on the left, and are paired on the right with the usual culprit for that process.

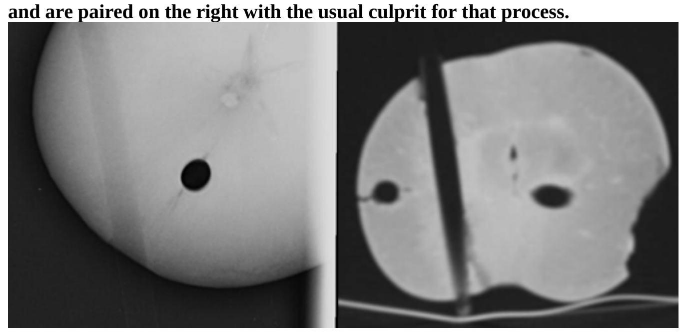

Fig. 3 (B) Experimental model of Air Bronchogram using the Apple/Straw setup. On the left is the plain film version showing one straw in profile (the air bronchogram), and one on end illustrating the potential for image variation depending on angle. On the right, the CT reconstruction, with the addition of a second straw in oblique (oval near midline) orientation.

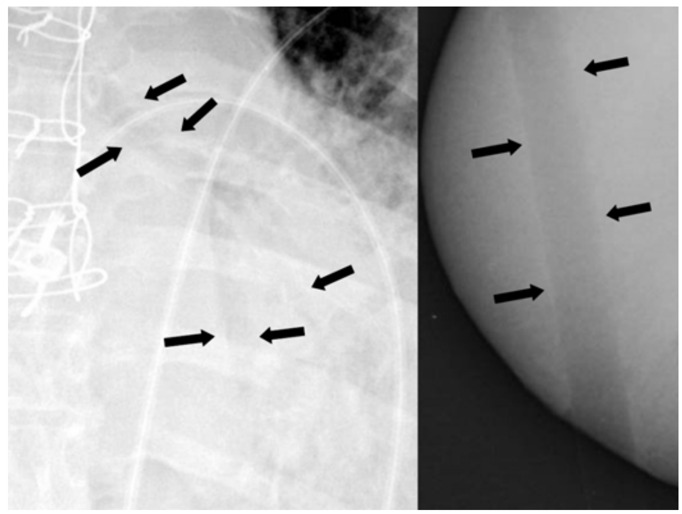

Fig. 4 (B) Side-by-side comparison of the Air-Bronchogram *in-vivo* (left) and using the experimental model (right). The arrows outline the patent airway as it courses through consolidated lung parenchyma (left). This process is analogous to the patent straw passing through an apple (right).

COPD, to include chronic bronchitis and emphysema, are acquired causes of airway obstruction and destruction and are most commonly accompanied by a long history of smoking.

Bronchiectasis, while often placed under the umbrella of COPD diseases, includes a broad range of disorders that deserve individual mention. By definition, bronchiectasis is the irreversible dilation of the bronchioles from any cause. Table 2 shows the three main categories of bronchiectasis. The underlying cause can be congenital, or secondary to severe infection, or secondary to an obstructive process. The most common infectious causes in adults are tuberculosis, methicilin-resistant *S. aureus.* In newborns, the TORCHES infections predominate: Toxoplasmosis, Rubella, CMV, Herpes, Syphilis, and others.

The first case has the most common cause of bronchiectasis, that is, Cystic Fibrosis. As with all causes of bronchiectasis, the initial insult is obstruction of the distal airways, setting the stage for infections and a release of the body's natural inflammatory mediators [4]. These mediators lead to further inflammation and destruction of the normal bronchiole architecture, leading to further dilation. As before, a cycle is created whereby dilated bronchioles are a site of recurrent infection, causing more inflammation and obstruction. Additionally, the underlying cause of the obstruction (i.e. increased mucus production, decreased ciliary motility, extrinsic compression, etc) continues to create obstructive/inflammatory sites.

Table 2: Causes of Bronchiectasis	
Congenital	Cystic Fibrosis
	Kartagener's Syndrome
	Alpha-1 Antitrypsin Deficiency
	Bronchomalacia
	Yellow-Nail Syndrome
Severe Inflammation	*Mycobacterium* (i.e. Tuberculosis)
	MRSA
	TORCHES
	Fungal Infection
Obstructive	Foreign Body Aspiration
	Bronchial Stricture
	Airway Mass/Tumor
	External Compression

Table. 2 Bronchiectasis involves a large differential diagnosis. For organizational purposes, it is useful to divide it into the 3 broad categories shown on the left. Within each category, the most common disease entities are shown on the right.

The processes involved in creating air-bronchograms and the associated differential is as extensive as the previous, but includes entirely different categories. As was discussed earlier, the features of air-bronchograms are a normal patent airway surrounded by lung tissue of increased radiographic density. It follows that the differential for air-bronchograms includes the differential for increased lung density and air-space processes. For organization purposes, it is helpful to start with three main categories: Consolidation, Atelectasis, and interstitial thickening.

Consolidation is by definition, a process of alveolar filling resulting in increased density on radiographs. The five common "sources" of a consolidation are hemorrhage, exudates, transudate, secretions, and malignancy. A useful mantra that is easily remembered is "Blood, Pus, Water, Protein, Cells." Table 3 contains the specific diagnoses for each of these categories.

Lower on the differential is atelectasis, the second source of air-bronchograms, and is defined as collapse of lung volume. There are five specific types of atelectasis: obstructive, compressive, cicatrization, post-operative and adhesive. Obstructive atelectasis occurs when circulating blood absorbs the gases in a section of lung that is no longer ventilated. The alveoli collapse, may fill with fluid, and are generally displaced by normal lung that expands to compensate.

Compressive atelectasis occurs secondary to loss of contact between the visceral and parietal pleura. In the upper lung, this is usually due to pneumothorax, and in the lower lung is usually due to pleural effusion. Less commonly, a pleural lesion or mass can cause impact on the neighboring lung tissue causing compression of the alveoli in that area. Cicatrization atelectasis is caused by scarring and fibrosis of the alveoli due either to long-standing infection (necrotizing pneumonia) or the effects of radiation exposure.

Table 3: Differential Diagnosis for Air Bronchograms		
Consolidation	Hemorrhage (Blood)	Embolism Trauma, contusion
	Exudate (Pus)	Pneumonia
	Transudate (Water)	Congestion, ARDS
	Secretions (Protein)	Mucus Plugs Alveolar Proteinosis
	Malignancy (Cells)	Alveolar Carcinoma Lymphoma
Atelectasis	Obstructive	
	Compressive	Pneumothorax Pleural Effusion Pleural Lesion
	Cicatrization	Radiation Severe Pneumonia
	Adhesive	Surfactant Deficiency
	Post-Operative	Low Tidal Volume
Interstitial Thickening	Linear Form (lines) *Remember "LIFE lines"*	Lymphangitic malignant spread
		Interstitial edema
		Fibrosis
		Edema
	Nodular (dots)	Granulomas
		Hematogenous Malignant spread
		Pneumoconiosis

Table. 3 Air Bronchograms can appear on radiographs in numerous processes. On the left are the three broad categories useful for organizing a differential. These can be subdivided into a general differential (middle column) and each of those has several possible specific etiologies (right).

Adhesive atelectasis is due to a surfactant deficiency and in adults is related to ARDS. Lastly, post-operative atelectasis is a common problem in thoracic and abdominal surgery. Diaphragm dysfunction, prolonged immobilization, and the effects of anesthesia all result in decreased tidal volumes and resulting collapse of under-ventilated alveoli.

The least common cause of air bronchograms is diffuse interstitial thickening and usually manifests as interstitial markings rather than air-bronchograms. The air-bronchogram is possible in these conditions as it is the surrounding lung architecture which is damaged, not the larger airways. If the interstitium thickens enough without being destroyed, an air-bronchogram will be visible. The potential processes involved are beyond the scope of this article, but Table 3 includes some of the more common diagnoses.

As this article has shown, two of the more common chest X-ray findings, tram-tracking and air bronchograms, can be confusing to identify and distinguish. With a thorough understanding of their pathophysiology and differential diagnoses, a variety of conditions and diseases can be either excluded or included based on a particular radiographs appearance.

The authors presented two cases with associated experimental models to help differentiate these two very different radiographic findings. An experimental model is also presented to help demonstrate how the 3-dimensional biologic activity translates to a 2-dimensional film.

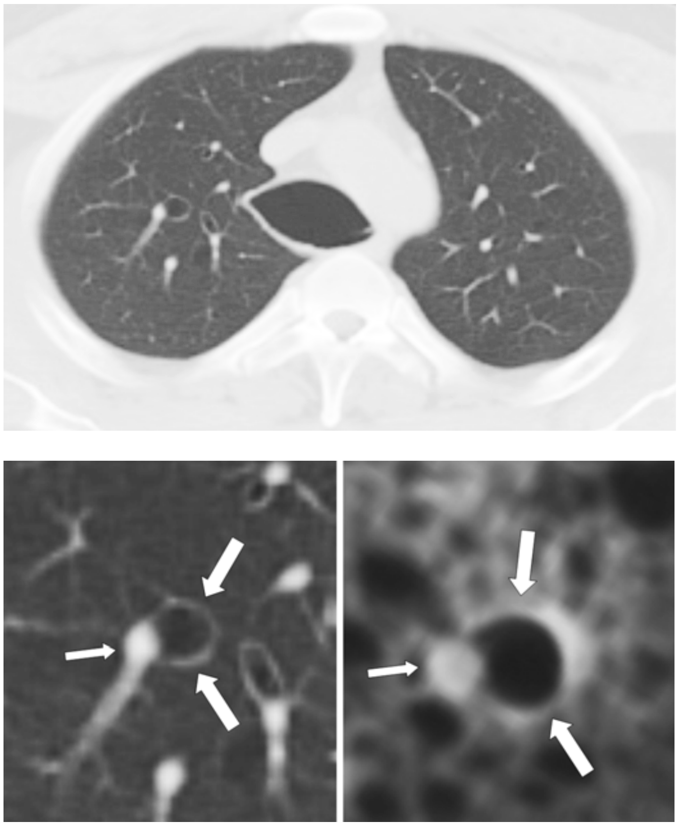

Fig. 5. Axial CT of the chest (upper and lower left) in a patient with bronchiectasis shows obvious signet ring signs, indicating bronchial dilation relative to the pulmonary arteries. The lower right image shows the experimental model image using the sponge. Two parallel straws of different diameters, the smaller having been filled with radio-opaque material to simulate adjacent vessel, the large straw with air to represent enlarged bronchiole.

Acknowledgements

The authors would like to graciously thank the following radiologic technologists for assisting with the experimental models: Raymond Fudge, RT; Ralph Alexander, RT; and Robert Robertson, RT.

Category 1 CME or CNE can be obtained on the MedPix™ digital teaching file on similar cases by opening the following link. Many Radiology Corner articles are also a MedPix™ Case of the Week where CME credits can be obtained.

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